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Case Report

Reversible ventricular dysfunction in cyanotic heart disease



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ABSTRACT

Ventricular dysfunction is a matter of concern for any preoperative cardiac patient. We describe 2 cases of cyanotic congenital heart disease (CCHD) awaiting on pump repair with hypoxia as a cause of ventricular dysfunction. Any Cyanotic Congenital heart disease presenting with ventricular dysfunction should be evaluated for treatable causes like hypoxia after exclusion of structural causes for the same.

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1. Introduction

Ventricular dysfunction in a preoperative patient is perceived a high risk factor for surgical correction of congenital heart diseases, particularly on cardiopulmonary bypass. It forms a very important factor for rejection of this subset for surgical correction. Hypoxia is a presumed cause of ventricular dysfunction in cyanotic heart disease with no reported literature on the same. We assume it to cause ventricular dysfunction but rarely attribute a ventricular dysfunction in cyanotic CHD to it. There is no case report of patient having improved ventricular function after treatment of hypoxia. We report 2 patients of cyanotic CHD who presented with ventricular dysfunction. Treatment of the same resulted in excellent immediate outcomes.

2. Case report

2.1. Case 1

A 17 year boy, with TOF underwent central shunt (Aorto-pulmonary shunt) at 5 years of age in view of hypoplastic branch pulmonary arteries. After the initial palliation he remained untreated and presented to us with severe cyanosis and dyspnea. Examination showed grade four clubbing and severe cyanosis (Saturation 55%). His hemoglobin was 22 g % and hematocrit was 71%. On examination, S1 was normal with single S2. There was a short ejection systolic murmur grade 2/6 at left upper sternal border. No continuous murmur was heard. Echocardiography showed Tetralogy of Fallot with hypoplastic pulmonary artery annulus of 14 mm (expected of

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20 mm). Right pulmonary artery (PA) was adequate at hilum (20 mm) and narrow at origin (12 mm), while left pulmonary artery was hypoplastic (8 mm against expected of 14 mm). There was tiny antegrade flow with no shunt flow seen. There was biventricular dysfunction (left ventricular ejection fraction, LVEF = 15–20%). A coronary cameral fistula from right coronary artery to right ventricle was also seen. Right atrium and right ventricle were not dilated. Patient underwent catheterization and angiography which showed similar findings. He underwent the investigations for possible causes of ventricular dysfunction (including serum calcium, metabolic workup which was all within normal limits). The possibility of hypoxia, coronary fistula leading to coronary steal and shunt induced ventricular dysfunction was discussed but no definite conclusion was arrived at. Initially he was rejected for surgical correction considering “very high risk”. However he later underwent Ventricular septal defect closure (Dacron patch) and right ventricle to pulmonary artery conduit (24 mm bovine pericardial tube with e-PTFE trileaflet valve) and division of thrombosed central shunt. The coronary cameral fistulae communication was considered small for any surgical intervention. His postoperative echocardiography at discharge revealed ventricular septal defect patch in situ, no residual shunt, well open right ventricular outflow tract, right ventricle-pulmonary artery conduit max PG = 15 mmHg, mild pulmonary regurgitation, Left ventricular ejection fraction (LVEF of 42%), showed marked improvement at the time of discharge. On follow up at 2 months the LVEF has normalized.

2.2. Case 2

A 7 day old female child born out of full term normal delivery with birth weight of 2.6 kg was referred for evaluation. Child at the time of presentation was asymptomatic with no cyanosis. Examination showed Normal S1 with single second heart sound and grade 3/6 ejection systolic murmur at left upper sterna border. Echocardiography showed her to be a case of Tetralogy of Fallot with borderline branch pulmonary arteries (right pulmonary artery was 3.6 against expected on 4 mm, left pulmonary artery was 3.4 mm), pulmonary annulus was hypoplastic. Since her baseline saturation was 94% it was decided to keep the child under regular. Child presented thereafter at 1 month of age in a very sick condition. She was acidotic (ph of 7.02), bicarbonate levels of 6 mg/ dl, with base deficit of – 23.2, tachypneic, cyanosed (saturation of 60%), she was pale with hemoglobin of 10 g/dl. Echocardiogram showed similar findings as the initial echocardiogram except there was severe infundibular stenosis with decreased antegrade flow. The left ventricular ejection fraction was 10%. Child required intubation at the time of presentation in view of acidotic breathing with poor respiratory efforts. The child was initially managed as cyanotic spell. Her saturations gradually improved to 85%. Child showed gradual improvement in ventricular function and was weaned off the ventilator by 8th day. Her ventricular function showed marked recovery and gradually improved to 25% by 11th day. Child was regularly followed on OPD basis. Her ventricular functions normalized over a period of 3 months and child underwent total correction at 5 months of age with a transannular patch with uneventful postoperative course.

3. Discussion

Ventricular dysfunction is a very important risk factor for mortality and morbidity in any cardiac surgery unit. Preoperative dysfunction is looked with skepticism by surgeons. Any child presenting with the same requires detailed workup for causes of ventricular dysfunction that includes looking for structural cause of ventricular dysfunction such as associated coronary involvement (including aberrant left coronary artery from pulmonary artery), left ventricular outflow tract obstruction and coarctation of aorta. In the setting of cyanotic CHD, which was a common factor in all of our patients, these are essentially rare. Shunt overflow over a long period may also lead to ventricular dysfunction as was thought for in our first case. The other cause prevalent in the general population is myocarditis/cardiomyopathy which is difficult to rule out in the absence of ventricular biopsies. Sustained ventricular and atrial arrhythmias can also lead to ventricular dysfunction (tachycardiomyopathy). None of the three patients discussed had any rhythm abnormalities. This manuscript elucidates the effect of hypoxia an entity scarcely recognized. While the first case who had continued untreated hypoxia for 17 years, demonstrates marked improvement in ventricular function after correction of hypoxia by total correction. In case 2 acute hypoxia, acidosis at the time of presentation could also have contributed to ventricular dysfunction. Ventricular dysfunction however persisted after the treatment of acidosis.

Although there has been multiple studies demonstrating sub cellular effects of chronic hypoxia on ventricular dysfunction, there have been no case reports on improvement of myocardial function after improvement of oxygenation.^{1,2} There have been a few animal studies reporting the effect of hypoxia but human data regarding the same in cyanotic CHD is lacking. Acute hypoxia such that occurs with severe prolonged spells or after cardiac arrest is known to cause ventricular dysfunction but improvement occurs very rapidly in these subset of patients after early resuscitation. Hypoxia could be a strong reversible risk factor for myocardial dysfunction especially in cyanotic (hypoxic) congenital heart disease. The hypoxic damage to the myocardium is further exaggerated by the altered rheological properties of the polycythemic state created by chronic hypoxia. Our case 1 patient who had outgrown the shunt (which got blocked too), presented at a very late stage. Although he adapted himself to chronic hypoxia by restricting his daily activity, the impact of chronic hypoxia was evident by the polycythemic status of the patient. Initial efforts to improve polycythemia by partial exchange transfusion did not give any benefit to the patient, while he waited for his surgery. We believe the correction of hypoxia by total correction relieved the important risk factor for myocardial dysfunction in the case.

4. Conclusion

Thus it is concluded that in cyanotic patients presenting with myocardial dysfunction reversible causes of cardiomyopathy like hypoxia, should also be considered after exclusion of

other structural causes of ventricular dysfunction. Correction of hypoxia by total correction of defect can lead to improvement in ventricular function in setting of cyanotic congenital heart disease.

Conflicts of interest

All authors have none to declare.

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